

## Fetus-in- Fetu Vs retroperitoneal mature teratoma: A revisit of the known rarity

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### Abstract

Fetus-in-fetu (FIF) is a rare pathological entity, not usually seen in the clinical practice. FIF is defined as the presence of one of the twins in the body of other. FIF may be controversial topic in its resemblance to retroperitoneal teratoma.

A-18-months old baby girl present with the abdominal mass. Radiological findings revealed heterogeneous retroperitoneal mass containing calcifications. Surgical exploration was done and the mass was successfully excised. Physical examination and histopathology of the mass confirmed the diagnosis of fetus-in-fetu.

FIF is a rare pathological condition and fewer than 200 cases have been reported worldwide with an incidence of 1 in 5,00,000 births. The comparative features of FIF vs retroperitoneal mature teratoma (RMT) and various theories of origin of FIF are discussed. The case is being reported in view of the unusual gross and microscopic findings and rarity of FIF.

**Keywords:** Fetus-in-fetu, Retroperitoneum, Teratoma.

### Introduction

Fetus-in-fetu is an extremely rare pathological condition with an estimated incidence of 1 in 5, 00,000 births<sup>1</sup>. Less than 200 cases have been reported worldwide<sup>2-4</sup>. Multiple theories are proposed regarding embryogenesis. To our knowledge, FIF was originally described by Meckel in 18<sup>th</sup> century.<sup>5</sup> Fetus-in-fetu, a term quoted by Willis, was described as rare condition in which a malformed parasitic twin was found inside the body of its partner usually in the abdominal cavity<sup>6</sup>.

Despite its prevalence among infants and children, has remained asymptomatic until later ages<sup>7</sup>. It is a fetiform calcified mass thought to arise from unequal division of the totipotent cell at blastocyst stage of embryogenesis give rise to a cellular mass parasitic within a mature embryo in a twin pregnancy<sup>8</sup>.

FIF can be differentiated from a retroperitoneal teratoma by calcified vertebral axis and limb buds within the mass<sup>9</sup>. We report a case of 18 months old baby girl presented with the abdominal mass. Final histopathological diagnosis of retroperitoneal FIF was rendered and differentiating points between mature teratoma were discussed.

### Case Report

A female child of 18 months presented with abdominal lump since birth which was gradual increased in size since 1 month with vague abdominal pain. The patient was born to 28 years old mother at thirty-nine weeks through normal delivery with prediagnosis at third trimester with abdominal mass. In view of painless, asymptomatic mass the patient was advised to wait and watch. She had been complaining of pain since 1 month now and in view of gradual increase in size, the patient was referred to Paediatric Surgeon. Upon physical examination, mass with a size of nearly 20x14x10cms was detected at left

hypochondrium and in lumbar region. The ultrasonography and computerised abdominal tomography showed a multilobated heterogeneous mass in the retroperitoneal area with calcifications. The tumour markers of the patient  $\beta$ -HCG and  $\alpha$ -FP were within normal limits. The haematological, biochemical and serological parameters were within normal limits. The provisional diagnosis of retroperitoneal teratoma vs. Fetus-in-fetu was made and exploratory laparotomy was done. The large variegated mass lying in the retroperitoneum measuring 19x15x11cms was removed in toto.(Fig. 1) and specimen sent for histopathological examination. The post-operative period was uneventful.



**Fig. 1: Intra-operative photograph of retroperitoneal mass as fetus-in-fetu**

**Gross Findings:** Received excised specimen of retroperitoneal mass composed of malformed foetus like structure comprising head, neck, abdomen and limb buds like structure in a large solid cystic mass.(Fig. 2) The mass measured 18x10x6cms and weighing 910 grams. External surface is sac like with partial skin covering at upper region and whole of the mass enclosed with thin smooth surface. The vertical cut section showed solid and large cystic areas with mucoid

fluid material exudes out. Upper pole of the mass showed hairs, pultaceous material, scalp and brain substance (Fig. 3). Middle segment showed vertebral column, cartilage, intestine and grey white solid areas (Fig. 4). Lower pole showed cyst with flattened wall and limb buds like structures attached to the external aspect. The gross specimen radiography was done and it showed bony densities of vertebral spine at cervical region (Fig. 5).



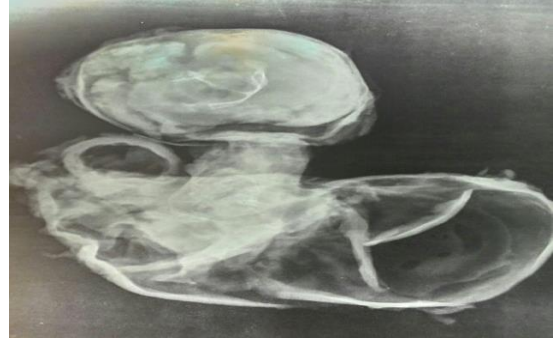
**Fig. 2: Gross photograph of malformed fetus like structure comprising head, neck, abdomen and limb buds like structure in a large solid cystic mass**



**Fig. 3: Cut section of upper pole of the mass showed hairs, pultaceous material, scalp and brain substance**

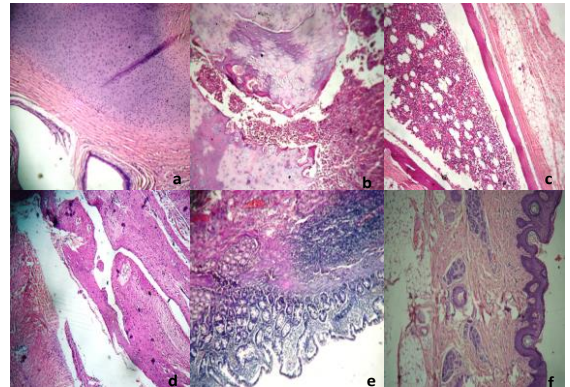


**Fig. 4: Middle segment showed vertebral column, cartilage, intestine and grey white solid areas**



**Fig. 5: Gross specimen radiography showed bony densities of vertebral spine at cervical region**

**Microscopic Finding:** Multiple sections from upper pole, middle segment and lower pole were taken. The histopathological structures identified and confirmed were glial tissue, cartilage, skin and adnexa and hairs. Middle segment reveals stomach, duodenum, small intestine, Vertebral column, bone and bone marrow, pancreas, soft tissue, lymph nodes, adipose tissue (Fig. 6). Lower pole showed cyst wall with flattened lining with no adnexal structures. There is no evidence of dysplastic or malignant component in the sections studied. Presence of vertebral column bone, cartilage, and other anatomical structures favours diagnosis of FIF over Teratoma. Final histopathological diagnosis given was fetus in situ.



**Fig. 6: Photomicrograph showing mainly components of FIF as cartilage, bone and marrow(a-c), glial tissue (d), intestine(e) and skin with adnexa(F) (H & E, x100)**

### Discussion

Fetus-in-fetu is extremely rare pathology to encounter roughly accounting one in 5,00,000 deliveries<sup>8</sup>, in which a malformed fetus is located in the body of its twin. FIF is rare condition with less than 200 cases reported in the lifetime<sup>2-4</sup>. Despite the detailed description in literature, its etiology and relationship with teratoma remained controversial. The exact embryogenesis of FIF is also controversial<sup>10</sup>.

Some investigators are proposing FIF occurs from the anomalous embryogenesis in a diamniotic

monozygotic twin pregnancy in which a malformed monozygotic twin lies within the body of its fellow twin<sup>11</sup>. Others consider it represent a highly organised teratoma<sup>10</sup>. FIF has a male preponderance with most of the patients presenting with an abdominal mass in 1<sup>st</sup> year of life. Other reported locations include the cranial cavity, the scrotum, sacrococcygeal region etc<sup>8</sup>. The number of fetus-in-fetu is usually single however; multiple foetuses-in-fetu have been reported<sup>12</sup>.

FIF was first coined by Meckel JF in the late 18<sup>th</sup> century to describe an encapsulated fetoid tumor within a fetus. In the literature fewer than 100 cases were reported. It must be differentiated from a retroperitoneal mature teratoma (RMT) which has an accumulation of pluripotant cells without organogenesis or vertebral segmentation<sup>5</sup>.

Over a period of time, the twinning theory is getting more famous<sup>13</sup>. Of late, Spencer has proposed that one or more of the following criteria's need to be

present to label any fetiform mass as FIF: i) Mass needs to be enclosed within a distinct sac, ii) Should be partially or completely covered by normal skin, iii) Should have grossly recognizable anatomic parts, iv) Should be attached to host by only few relatively large blood vessels, v) Either be located immediately adjacent to one of the sites of attachments of conjoint twins or associated with neural tube or the gastrointestinal tract<sup>14</sup>.

In our case, both the criteria of Willis and Spencer are fulfilled and finally diagnosed as FIF. Differential diagnosis of FIF is retroperitoneal masses, such as Neuroblastoma, Wilms tumor, Teratomas and hydronephrosis etc. The main diagnostic challenge lies in differentiating FIF from the teratoma, as it may undergo carcinomatous changes. But the final important feature that has been used to distinguish between FIF and teratoma is the presence of the vertebral column<sup>9</sup>.

The various differences were enlisted in Table 1.

**Table 1: Differences between FIF and RMT<sup>9</sup>**

Description	Fetus-in-Fetu (FIF)	Retroperitoneal mature Teratoma (RMT)
Definition	"A vertebrate fetus included within the body of its partner.	"A true tumour or neoplasm composed of tissue from multiple germ layers.
Size--	Usually small	Large
Presence of capsule	Yes, often containing a small amount of fluid simulating an amniotic sac	Characteristically does not have a capsule
Suspending Pedicle	Vascular Pedicle suspends the FIF within the capsule though no direct connection between these and the host vessels may be found	No pedicle, just a broad attachment to the posterior abdominal wall
Well-formed fetal parts	Usually present. Well-formed limbs are often seen, but their ratio to trunk is way behind the normal expected. Mostly acardiac and anencephalic	May be seen once in a while like single digit, teeth in bony sockets, intestinal mucosa, respiratory mucosa, cartilage.
Segmented vertebral axis	Presence is pathognomonic of the diagnosis	Never seen.
Growth	Initially parallels that of its twin, but ceases subsequently either due to an inherent defect or vascular dominance of host twin and may actually retrogress	Present in fetal life and continues to grow progressively
Malignant change/ recurrence	Rare	Definite malignant potential if left untreated
Workup for secondaries	Not required.	Always required
Surgical excision in an infant	Not an emergency unless there are symptoms of compression	Infant should be operated as early as possible since delay can lead to malignant transformation.

### Pathological aspect

Detailed and meticulous gross as well as histopathological examination of the excised specimen is the "gold standard" for diagnosing FIF. The most common seen organs are vertebral column limbs and CNS followed by gastrointestinal tract, vessels and urinary tract. Most of these histopathological structures with vertebral column are found in our case.

Complete surgical excision is the treatment of choice in FIF to prevent complications like infections, infarction or spontaneous haemorrhage and recurrence when the membrane is left behind<sup>9</sup>.

## Conclusion

The case presented in our report meets all the accepted criteria of abdominal FIF. Fetus-in-fetu is a rare, interesting medical curiosity that typically presents with abdominal mass in early childhood. FIF is considered as a benign condition, while the potentially malignant characteristics of teratoma constitute the basis of the discussion. Complete excision in toto is curative and allows confirmation of diagnosis by histopathology. Pathologically, presence of vertebral column is used to distinguish between FIF and RMT. Future research efforts should be made to establish the true nature of FIF.

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